

# STATES RELATED TO OR CONFUSED WITH COMA

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Neurologists are increasingly asked to see patients with acute neurological problems, many of whom have disorders of consciousness and awareness. The intensive care unit is a common source of referral of such patients where common questions include:

- ▶ Why isn't the patient waking up?
- ▶ Is the patient going to recover?
- ▶ Is the patient brain dead?

The neurologist in training needs to have a clear understanding of the basis of normal consciousness, to be aware of the range of disorders of consciousness, and particularly to be aware of the pitfalls in diagnosis.

## CONSCIOUSNESS

Consciousness is a state characterised by awareness of self and environment and an ability to respond to environmental factors. Normal consciousness can be regarded as having two separate but closely interrelated components. The first of these is the arousal component of wakefulness. It is this that keeps the patient awake and which relates to the physical manifestations of awakening from sleep—for example, eyes being open, motor activity. The second component is the content of consciousness or the awareness of self and environment. This consists of the sum of psychological functions of sensations, emotions, and thoughts.

A detailed description of the pathophysiology of consciousness is beyond the scope of this article. In simple terms, normal consciousness requires an interaction between the reticular activating system of the brain stem and the cerebral cortex. The reticular activating system is responsible for arousal or alertness and the cerebral cortex is responsible for the content of consciousness or the awareness of self and environment.

It should be recognised that an individual's awareness of self can only be recognised by an observer on the basis of the responses made to external stimuli, especially verbal or motor responses. Inability to respond may not only result from abnormalities of consciousness but also from paralysis (such as the locked in state). This may be a particular problem in the intensive care unit where patients have often received sedative drugs and paralytic agents.

Issues relating to the common patterns of altered consciousness, sleepiness, delirium, and coma are dealt with elsewhere in this supplement. This article will introduce some of the less commonly seen states.

## STUPOR

In this state the individual appears to be asleep and yet, when vigorously stimulated, may become alert as manifest by eye opening and ocular movement. Other motor activities are limited and there is usually no speech.

Although a proportion of patients in stupor have diffuse organic cerebral dysfunction, a variety of psychiatric disorders produce an identical clinical picture.<sup>1,2</sup> It may be difficult to differentiate stupor resulting from organic cerebral dysfunction from those with catatonic schizophrenia or severe depression. The EEG is usually of considerable help as in organic stupor it shows a diffuse abnormality whereas in other forms of stupor it is usually normal. The exception is in patients with catatonic stupor or catatonia.<sup>3</sup>

Caloric testing in organic stupor will usually reveal tonic deviation, whereas in psychiatric stupor ocular nystagmus will be present. This is because the fast phase reflects the correction following tonic deviation and this requires the patient to be conscious.

## AKINETIC MUTISM

Akinetic mutism was first described in patients thought to have suffered diencephalic damage.<sup>4</sup> The syndrome is characterised by immobility and eye closure with little or no vocalisation. Sleep/wake cycles can be seen, as indicated by eye opening. There is little in the way of movement to

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painful stimuli and the hallmark is the relative paucity of signs indicating damage to descending motor pathways, despite the immobile state. In other words spasticity and rigidity are not usually evident. The best presently available clinical and pathological evidence indicates that akinetic mutism can arise as a result of lesions that interfere with reticular cortical/integration but spare the corticospinal pathways. There is some debate about whether or not the syndrome should be clearly differentiated from the vegetative state. A patient in the fully established vegetative state will almost invariably show spasticity and rigidity of the limbs which are absent from patients with the syndrome of akinetic mutism. In the early stages of the vegetative state, the two clinical syndromes are indistinguishable.

### Vegetative state

This was the term suggested by Jennett and Plum<sup>5</sup> to describe patients who recover the arousal component of consciousness but not, as far as can be determined, awareness. The clinical picture is that of a patient who shows eye opening but who has no voluntary movement and who is unaware of self and environment. There is often evidence for sleep/wake cycles with either complete or partial preservation of brain stem functions.

The clinical picture may be seen as a transient phase during the recovery of acute brain damage or may be permanent as a consequence of the failure to recover from brain damage. The same clinical picture may be seen as the result of the progression of a degenerative neurological disorder such as Alzheimer's disease. The Royal College of Physicians produced a series of clinical criteria.<sup>6</sup> (1) There should be no evidence of awareness of self or environment at any time. There should be no volitional response to visual auditory, tactile or noxious stimuli. There should be no evidence of language comprehension or expression. (2) There should be presence of cycles of eye closure and eye opening which may simulate sleep and waking. (3) There should be sufficiently preserved hypothalamic and brain stem function to ensure the maintenance of respiration and circulation.

There is some debate about the use of the terms permanent and persistent in relation to the vegetative state. The permanent vegetative state should be diagnosed when there can be a high degree of certainty of irreversibility (see prognosis).<sup>7</sup>

There have been claims that patients who were thought to be in the persistent vegetative state may occasionally make a late recovery with some volitional movement indicating awareness.<sup>8</sup> The evidence suggests that such cases are excessively rare if careful and repeated observations are made. It is particularly important to assess whether any movements made by a patient thought to be in the vegetative state are reflex or under voluntary control. In the vegetative state eye blinks are reflex, whereas in the locked in syndrome, the eye blinks are under voluntary control and the patient can blink to command.

The pathological basis of the persistent vegetative state is usually that of widespread cortical damage resulting from such causes as cerebral hypoxia or widespread subcortical damage resulting from severe head injury.

Many terms over the years have been used to describe patients with a similar clinical picture. These include coma vigil, apallic syndrome, cerebral death, near cortical death, and total dementia.

### Locked-in syndrome

The ventral pontine or locked in syndrome describes a condition of total paralysis below the level of the third nerve nuclei. Such patients can open their eyes and elevate and depress their eyes to command. Horizontal eye movements are usually lost and no other voluntary movement is possible. The diagnosis of this state depends on the recognition that the patient can open his eyes voluntarily rather than spontaneously in the vegetative state.

The neuropathological basis for this condition is usually infarction of the ventral pons and efferent motor tracks. A similar clinical picture may sometimes be seen in patients with pontine tumours, pontine haemorrhage, central pontine myelinolysis, head injury or brain stem encephalitis. Recovery is exceptional.<sup>9</sup>

### Catatonia

This state is usually associated with psychiatric illness (affective more than psychotic) but may occur in metabolic and drug induced disorders. Patients do not move spontaneously, seem unresponsive to their surroundings, but appear conscious. Neurological examination is normal. The catatonic state is characterised by eyes open and unblinking, pupils dilated but reactive, oculocephalic responses absent or impaired, and caloric responses intact. Passive limb movements are met with a "waxy flexibility". This state is difficult to distinguish from organic disease, particularly in lethargic unresponsive individuals. The electroencephalogram (EEG) shows a low voltage fast record rather than the "slowing" of true coma.

### Psychogenic coma

In psychogenic coma the eyelids are kept firmly shut and are resistant to opening. Oculocephalic responses are unpredictable though nystagmus is evident on caloric testing. Motor tone is normal or inconsistent and limb reflexes retained. Other physical signs based on reflex self protection have been used in this syndrome though their validity has not been formally assessed.

The EEG shows awake rhythms.

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